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# Xeroderma Pigmentosa a Dermatological Disease and Potential Exacerbating Effects of Anaesthesia

**Lalwani P*, Pandey R and Kashyap L**

Department of Anaesthesiology, Critical care Pain and Perioperative medicine. All India Institute of Medical Sciences, New Delhi

***Corresponding author:** Lalwani P, Department of Anaesthesiology, Critical care Pain and Perioperative medicine. All India Institute of Medical Sciences, New Delhi

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## Abstract

Xeroderma Pigmentosa (XP) is an autosomal recessive disorder, affected patients are extremely sensitive to ultra violet (UV) rays and have characteristic lesions in sun exposed areas viz head, face and neck. 25-30% of XP individuals develop neurological manifestations in infancy or second decade. In vitro studies and case reports suggests worsening of neurological status in XP patients after exposure to inhalational agents. We present successful anaesthetic management of a young boy presented to us for excision of squamous cell carcinoma of right eye lower lid. Perioperatively patient was covered and kept in dimlight. Anaesthesia was induced with fentanyl, propofol, atracurium and maintained with oxygen, air and sevoflurane. Shifted to dimlight ward with goggles to avoid photophobia and discharged after three days. In our case sevoflurane did not cause any adverse outcome. Further research is needed in this group of patients to know the effect of anaesthetic agents in worsening the disease course as these patients are frequently exposed to anaesthesia for various surgical lesions.

## Keywords

Ultraviolet radiation; Halogenated agents neurodegeneration

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## Introduction

Xeroderma pigmentosa (XP) is a rare autosomal recessive disorder with an incidence of 1 in 2.5 million people in USA 1:20,000 in Japan [1].Incidences in Northern African and Western Asian countries, such as Libya, Tunisia, Morocco, and Pakistan, may be higher due to more frequent [2]. Affected patients are often referred as night or moon people as they are extremely sensitive to sunlight and ultraviolet rays and have characteristic lesions in sun exposed areas viz head, face and neck. Apart from dermatological lesions these patients also suffer from ocular and neurological symptoms and have 1000 times higher chances of malignancies at an early age especially in sun exposed areas [3]. We present anaesthetic management of a young boy presented to us for excision of squamous cell carcinoma of right eye lower lid.

## Case Report

An eight year old boy weighing 20 kg presented to our hospital with XP with right eye lower lid squamous cell carcinoma (1.5 x 1 cm). He was posted for excisional biopsy of the mass. At the age of 2 years child developed skin lesions at face which spread gradually to entire face and extremities. Between the ages of 5-7 years he had history of basal cell carcinoma with small ulcero infiltrative lesion of medial canthus of left eye, left lower lid, root and tip of nose and of scalp. For which he underwent electrosurgical excision, electrochemotherapy with bleomycin and electrofulguration under general anaesthesia (GA) three times. Recently 2 months back he has received radiotherapy for mass lesion over left eye that resolved after treatment (Figure 1).

**Figure 1:** Radiotherapy for mass lesion over left eye.

On examination he had marked freckled pigmentation all over the face trunk and extremities and multiple scar marks over face (Figure 2).

**Figure 2:** Freckled pigmentation over the face and trunk.

Airway examination revealed adequate mouth opening with healed oral ulcers which developed because of recent radiotherapy. His neck movements were normal. Examination of right eye revealed corneal opacity with nasal neovascular tissue and diminished distant vision. Rest of the physical examination and investigations were within normal limits. He was on tab isotretinoin 10 mg OD and fucidin cream since 4 months.

Patient was fully covered and kept in dimlight in preop room. EMLA cream applied over the dorsum of the hand. Inside the operation theatre (OT) patient was covered fully, routine monitors attatched ECG, NIBP and pulse oximetry. Intravenous cannulation done and anaesthesia was induced with injection fentanyl 40 mcg, propofol 40mg,atracurium 10 mg and proseal LMA size 2.5placed in a single attempt.

Anaesthesia was maintained with oxygen, air, sevoflurane and fentanyl top up 20 mcg. Injection paracetamol 375 mg given for analgesia and ondansetron 3mg given 15 minutes before end of surgery. Surgery lasted for 2 hours. Patient reversed and extubated at the end of surgery. He was shifted in the ward with dim light with dark goggles to avoid phtophobia and observed for three days .He had an uneventful recovery and was discharged with advice of regular follow up.

## Discussion

Xeroderma pigmentosa is characterized by increased propensity to DNA damaging effects of ultraviolet radiation (UVR). Eight different genes XP-A to XP –Ginvolved in neucleotide excision and repair are affected. Disease typically starts with dermatological symptoms at preschool age and present invariably in all patients suffering from XP. Skin manifestations comprises of severe sunburn in sunexposed area, persistent erythema, freckle like pigmentation, dry pigmented skin, atrophy, keratosis and telangiectasis. Intravenous canulation and fixing may be difficult because of freckling and dryness. These patients may have difficult mask ventilation, laryngoscopy & intubation because of ulceroproliferative mass on face, restricted mouth opening, dryness, cheilitis, oral ulcers & scarring. Nearly 80-90% of XP patients suffer from ocular pathology as anterior parts of the eye are exposed to UVR.

Most common symptoms are photophobia, conjunctivitis, corneal neovascularization, dry eye corneal scarring, ectropion, blepharitis conjuctival melanosis and cataract. Eyelids and surfaces of the eye may

be severely affected with 2000 fold increased risk of malignancy of eye and surrounding ocular tissue. Central nervous system does not have direct UV radiation exposure but significant subset of XP patients18-24% suffer from neurological problems .Unrepaired oxidative damage may be possible mechanism of neurodegeneration which include loss of intellectual functioning, impaired hearing abnormal speech, areflexia, ataxia, peripheral neuropathy and loss of ability to walk and talk. 25-30% of XP individuals develop neurological manifestations in infancy or second decade.

Imaging studies and pathological examination suggest neuronal loss, cortical atrophy and ventricular dilatation without inflammation. Patients with XP-D and XP-A mutation are most affected with neurological symptoms, while XP-C, XP-E and XP variant do not have neurological disorder [4].

Halothane, isoflurane and sevoflurane effect neucleotide excision repair and may worsen neurological symptoms. Fjauji reported postoperative neurological aggravation after sevoflurane anaesthesia in an adult patient already suffering from neurological dysfunction [5]. Hajijafari, Webb used sevofiurane without any untoward complication [6,7]. Reitz and Lanz showed deoxyriboneucleic acid (DNA) strand breaks after in vitro exposure to halothane and suspected its genotoxic side effect in XP patients [8].

We used sevoflurane and observed our patient for 3 days in the ward he had an uneventful recovery. Nitrous oxide should also be avoided to prevent myelosuppression as these patients are frequently prescribed 5-fluorouracil. TIVA with propfol, ramifentanil is recommended and dexmedetomidine has been used successfully. Increased sensitivity to benzodiazepines, opioids and muscle relaxants has been reported in XP patients.

Oliveria et al recommends minimal use of muscle relaxants with neuromuscular monitoring [9]. These patients may be on treatment with chemotherapy and radiotherapy because of high incidence of cancers. Side effects of these treatments need consideration, our patient received bleomycin and had radiothearpy induced healed oral ulcers. Lack of sunexposure causes vitamin D deficiency osteoportic bones and immunedeficiency implies careful positioning during perioperative period.

Perioperatively patients needs to be protected from UVR. Surgical headlamps and operating room spotlights emit UVR 10-14$\mu W/cm^2$ and exposure to UV R $>2\ \mu W/cm^2$ should be avoided. Open surgery is preferred over laparoscopic surgeries as UVR is emmited from laparoscopic light source. Vinyl UV – filtering film can be used to cover OT spot lights and patient should be properly covered and kept in dim light [7].

As we have a successful outcome even after using sevoflurane, more clinical and experimental studies are required to affirm and explain neurotoxicity of halogenated agents in this group of patients.

## Conclusion

XP is primarily a dermatological disease with predisposition to various skin cancers and unusual perioperative implications. More research is needed on perioperative management of XP patients as their number is considerably high and they require frequent surgeries.

## References


1. MulimanI SM, Talikoti DG. (2013) A child with xeroderma pigmentosum for excision of basal cell carcinoma. Saudi J Anaesth. 7(4):467-69.
2. Bhutto AM, Shaikh A, Nonaka S. (2005) Incidence of xeroderma pigmentosum in Larkana, Pakistan: a 7-year study.Br J Dermatol. 152(3):545-51.
3. Lehmann AR, McGibbon D, Stefanini M (2011) Xeroderma pigmentosum. Orphanet J Rare Dis. 6:70. https://rarediseases.org/gard-rare-disease/xeroderma-pigmentosum/
4. Fjouji S, Bensghir M, Yafat B, Bouhabba N, Boutayeb E, et al. (2013) Postoperative neurological aggravation after anesthesia with sevoflurane in a patient with xeroderma pigmentosum: a case report. J Med Case Rep. 7(1):73. Hajijafari M, Ziloochi MH, Fazel MR. (2014) Inhalation anesthesia in a patient with xeroderma pigmentosum: a case report. Anesth. Pain Med. 4(3):e17880.
5. Webb LM, Chatterjee D, Brockel MA. (2021) Perioperative management of a patient with Xeroderma Pigmentosum: A case report. J. Pediatr. Surg. 64(2): 101710.
6. Reitz M, Lanz E. (1993) DNA strand breaks in cells with DNA repair deficiency after halothane exposure in vitro. Arzneimittel-Forschung. 43(4):418-20.
7. Oliveira CR, Elias L, Barros AC, Conceição DB. (2003) Anesthesia in patient with Xeroderma Pigmentosum: case report. Rev Bras Anestesiol. 53(1):46-51.



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